ARTICLE III.

ON CONGENITAL ATRESIA OF THE PULMONARY ARTERY.[1] By ALBERT F. STIFEL, M.D., of Wheeling, West Virginia.

CASE.—A female infant was brought to the St. Jacobs Hospital in Leipzig for treatment of a blennorrhœa conjunctivæ. In the hospital record nothing is mentioned of cyanosis, which was probably absent. The infant died when six weeks old from exhaustion, without any particular premonitory symptoms.

Post-mortem examination[2] 15 hours after death.—Poorly nourished; eyelids adherent; conjunctivæ covered with a mucous substance; umbilicus well healed; brain engorged with blood of a deep purple colour; the greater part of the lungs distended with air, but some atelectatic spots of a blue colour present. The heart, altogether, large; greatest length, 48 mm., greatest breadth 35 mm. The walls of the right ventricle are very thick, measuring at the base 6 mm., at the apex 9 mm., but they surround a very small sinus. The walls grow thinner towards the infundibular portion, but still remain of considerable thickness. On trying to pass a probe from the infundibular portion of the right ventricle to the pulmonary artery, an obstacle is found, which, on closer examination, proves to be a membranous septum, entirely obliterating the orifice of the pulmonary artery. The position of this septum corresponds to the semilunar valves, which appear to have grown together. Beyond this septum the pulmonary artery is permeable, and communicates with the aorta through the open ductus arteriosus. The circumference, including the walls, of the aorta before meeting the ductus arteriosus is 30 mm., the pulmonary artery 17 mm., the ductus arteriosus 9 mm. In the septum of the ventricles, corresponding to the pars membranacea, there is an aperture measuring 3.5 mm. In the right ventricle it is between the anterior and left tricuspid, in the left ventricle between the posterior and right mitral valve. The tricuspid valves are normal. The foramen ovale is open, only a few fibres of the endocardium passing over it; largest diameter 10 mm., smallest 6 mm. On the left side of the heart the ventricle appears to be considerably dilated. The walls measure at the base 4 mm., at the apex 5 mm. in thickness. The valves of the aorta are normal, while on the anterior mitral valve there is a rugged excrescence. The course of the aorta and pulmonary artery, with the above-mentioned exceptions is normal. Both kidneys are very small; the one measuring: length 17 mm., breadth 10 mm., thickness 5 mm.; the other: length 20 mm., breadth 11 mm., thickness 4 mm. The cortical substance is very thin. On the surface of one kidney there are small cysts containing a clear liquid. Microscopical examination proved the existence of an interstitial nephritis. Otherwise nothing abnormal could be found at the urino-genital organs. The spleen is large. The intestinal tract, with the exception of a Meckel's diverticulum, normal. The liver is small, firm, and of a dark purple colour. The umbilical arteries have a knotty appearance, and contain some dark red thrombi.

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[1] An Abstract of an Inaugural Dissertation presented in 1879 to the Medical Faculty of the University of Leipzig, for the degree of M.D.

[2] By Prof. Cohnheim, through whose kindness I am permitted to use this case.

Of the 46 cases of atresia of the pulmonary artery which I have found described in the literature of the subject, the trunk of this artery was represented by a solid cord in six cases; in thirty cases the pulmonary orifice was completely closed, while in the ten remaining cases it could not be ascertained exactly where the obliteration had taken place. In thirty-five of these cases the septum ventriculorum had an aperture, the defect usually being near the base. The ductus arteriosus was permeable in thirty-eight cases, closed in four. The foramen ovale was found open in twenty-eight cases, closed in four, not recorded in fourteen. Twenty-three died before the close of the third month, eleven between the third month and end of twelfth month, seven between the first and twelfth year, one reached the age of twenty-one years, another thirty-seven years, three unknown. In eleven cases the orifice of the aorta was moved to the right, above the septum of the ventricles, or, as in some cases, to the right of the septum, arising completely from the right ventricle.

When the pulmonary artery is closed the blood of the right side can flow to that of the left by passing through one or both of the apertures in the septum of the auricles or that of the ventricles. Either one of these is quite necessary for the circulation of the blood. After the blood has entered the left side of the heart there is no further hindrance to its re-entering the systemic circulation. The pulmonic circulation receives its blood from the aorta through the ductus arteriosus. In four cases the ductus arteriosus has been found either absent or closed. In the extraordinary case of Voss,[1] where the man reached the age of thirty-seven years, the bronchial and the anterior coronary arteries were found very large, permitting a sufficient pulmonary circulation for so many years. In the case of Mallwo[2] the bronchial arteries were not examined. Also in a case of Fearn,[3] the means by which the lungs were supplied with blood was not ascertained. In the case of Chevers[4] the left subclavian artery is supposed to have supplied the lungs with blood. With an open ductus arteriosus and an aperture in the septum of the auricles or ventricles the atresia of the pulmonary artery will be no hindrance to the circulation, provided all those openings are large enough. An imperfect arterialization of the blood will be a necessary result of this deformity in any case. The venous blood of the right side is mixed with the arterialized blood of left auricle, which comes from the lungs, to pass through the left ventricle to the aorta; partly to pass into the systemic circulation, but also to enter the pulmonic circulation through the ductus arteriosus. Thus, while the systemic circulation receives some venous blood, part of the arterialized blood which has just passed through the pulmonic circulation is returned to it again.

It can hardly be doubted that these deformities of the heart are congenital. The death at an early age, the symptoms in many cases of imperfect arterialization of the blood from a time soon after birth, the open ductus arteriosus and foramen ovale, and especially those cases where death has taken place during the first few days of life, leave no room for doubt on this point. The septum ventriculorum grows from the apex towards the base of the heart, and this part is the last to completely separate the two ventricles. Embryologists agree that this septum is completed between the sixth and eighth week of intra-uterine life. If the development of this septum is retarded by the blood, which has no other way of escape from the right ventricle when the pulmonic orifice is closed than to pass through the aperture of the undeveloped septum to the left ventricle, then the impediment to the escape of blood in the pulmonary artery must have existed before the close of the second month. Meyer,[1] Peacock,[2] Kussmaul,[3] and nearly all the writers on this subject believe that this.septum is retarded in its development and not perforated by a pathological process as v. Dusch[4] and Bouillaud[5] have claimed. If the theory of perforation of the septum were correct, then we would expect to find the edges of the aperture showing a pathological process in many cases, we would expect a perforation of the septum to occur during after-life; and unless the septum of the embryo should be peculiarly liable to perforation, we would expect to find a perforation of the septum, both in intra- and in extra-uterine life, without any other alterations of the heart. But the contrary is usually found: the edges of the aperture, as in our case, are nearly always smooth, and perforation without any other abnormality is a very rare occurrence. That the pressure of the blood in the right ventricle cannot perforate a once completed septum, is shown in those cases of atresia of the pulmonic orifice in which the septum of the ventricles was found complete, the blood escaping through the foramen ovale and the right ventricle being a useless appendage. Such cases have been described by Hunter,[6] Rokitansky,[7] Hare,[8] Peacock,[9] Olivier,[10] and others.

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[1] Norsk. Magazin., 1857.

[2] Virchow's Archiv, Bd. XIX., p. 438.

[3] See Peacock on Malformations of the Heart. 2d Edition, London, p. 74.

[4] Ibid.

The majority of writers on the subject agree that the obstruction in the pulmonary artery is the primary change, which causes the foramen ovale and ductus arteriosus to remain open and hinders the perfection of the septum of the ventricles. If this obstruction comes before the completion of the septum ventriculorum, then the further development of the septum will be deterred. If the septum is complete the blood will have to pass through the foramen ovale, which current will cause the foramen to remain patent.

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[1] Virchow's Archiv, Bd. XII. p. 518.
[2] Loc. cit.
[3] Zeitschrift für rationelle Medecin, Bd. XXVI. p. 118.
[4] Verhandl. des naturhistorich-medic. Vereins zu Heidelberg, Bd. I.
[5] See Peacock, loc. cit., p. 162.
[6] Med. Observ. and Inquir., vol. iv. p. 330.
[7] Wochenblatt d. Zeitschrift der k. k. Gesellschaft der Aertzte z. Wien. I. No. 14.
[8] Peacock, loc. cit., p. 71.
[9] Idem, p. 72. Case VI.
[10] Bull. de la Soc. Anat., vol. xxxvi. p. 320.

Should the aperture of the septum ventriculorum not be large enough then the blood will accumulate in the right auricle and flow through the foramen ovale, keeping both passages open for the escape of the blood of the right side. If the lungs receive a small quantity of blood through a small ductus arteriosus, the left auricle will also receive less, and the weaker pressure in this auricle, as compared with that of the right, may assist in causing a current through the foramen ovale and keeping it open. The unequal pressure in the aorta and pulmonary artery will cause the blood to flow through the ductus arteriosus; the current will then prevent its closing. The expansion of the lungs in after-life, which will cause the aspiration of blood as well as air, will help to lower the pressure in the pulmonary artery and will make the difference in pressure still greater. In those cases where the orifice of the aorta appears to be moved to the right and to arise partly or wholly from the right ventricle, Meyer believes that a dilatation of the right ventricle has pushed the septum of the ventricles to the left, so that its free margin comes to stand under or to the left of the aorta. Heine,[1] having described a case of this kind, believes that the deviation of this septum to the left is a primary malformation, causing, as he concludes, the atresia of the pulmonic orifice. He thinks that if the aorta arises from the right ventricle the pulmonary artery will become useless and obliterated. But, as Kussmaul has already shown, we cannot conceive any position of the pulmonary artery and aorta in the right ventricle, in which blood will not be pressed into the one as well as the other. A current will pass through both, and either current will not stop for a teleological reason alone. Kussmaul believes that the obstruction in the pulmonary artery before the septum of the truncus arteriosus meets with that of the ventricles prevents the semispiral turn, which the pulmonary artery and aorta normally make, causing the aorta to arise further to the right than usual.

Halbertsma[2] not only agrees with Heine, in so far that he believes the septum ventriculorum deviates in these cases by perversion in the process of development, but he claims that the septum of the truncus arteriosus also deviates from a more or less vertical direction to take a more horizontal course, until its lower margin combines with what would be the pulmonic orifice and obstructs it, instead of connecting with the septum of the ventricles. I think Halbertsma has overlooked one obstacle to the deviation of the septum of the truncus arteriosus. As soon as this septum turns to take a horizontal position, and closes the pulmonic orifice, a great part of the truncus arteriosus will be occluded, and the aorta will then have to carry a current equal to that carried by the whole truncus arteriosus heretofore. If a current of blood can prevent the closing of the ductus arteriosus and foramen ovale, which it certainly can, then it is highly probable that such a current, by its pressure on the septum of the truncus arteriosus will prevent this from deviating to a horizontal direction and obstructing about one-half of the truncus arteriosus. An inflammation or any other pathological process will have to overcome the same force, but we have ample proof that a pathological process can overcome a very strong current in the constriction of the orifice of the aorta in after-life, where a complete atresia would probably result in many cases if life would not necessarily cease before that time. In a case which Halbertsma describes, the longest diameter of the right auriculo-ventricular aperture was parallel to a frontal section instead of the usual nearly sagittal direction. He claims this could only be explained by a perversion of the process of development or deviation of the septum ventriculorum, which drew the anterior corner of the aperture to the left. It may be that the septum draws the anterior corner of the aperture to the left, but why should this only be possible by a perversion of the process of development and not by a septum which is pushed to the left during a dilatation of the right ventricle?

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[1] Angeborene Atresia des Ostium arteriosum dext. Dissert., Tübingen, 1861.

[2] Archiv f. d. holländ Beiträge, III. p. 387.

Equally interesting and important is the question why the valves and apertures of the right ventricle are the favourite seat of disease during intra-uterine life, while in after-life those of the left side are, in nearly all cases, the first to be affected, yet a satisfactory answer has not been given. Friedreich[1] thought the walls of the right ventricle had to overcome a greater pressure during the systole of the heart in fœtal life, while in after-life it is known that the left ventricle has the greatest pressure to bear. He concluded that the ventricle which bears the greater pressure and friction is the most liable to disease. However plausible this explanation may seem for the predisposition of the left ventricle in after-life to inflammation, it will not be so satisfactory in those cases of congenital atresia of the pulmonic orifice where the septum of the ventricles is not completed. In these cases the foramen ovale permits the equal division of blood for the two ventricles, and before the septum of the ventricles is completed, or when only a rudiment of such is present, as in some cases, it is difficult to understand how one side of these united ventricles should have to exert more force, bear more pressure, or be subject to more friction than the other. If the thickness of the ventricular walls of the fœtal heart show the relative proportion of pressure the respective ventricle has to bear and overcome, then, indeed, it would appear that the left ventricle does the most work before its septum combines with that of the truncus arteriosus. Kölliker, in his *Entwickelungsgeschichte*, 2d ed., page 909, fig. 557–3, gives an illustration of a section of the heart of a four weeks' old embryo, with a rudimentary septum ventriculorum, in which the walls of the left ventricle are considerably thicker than those of the right, and Kölliker distinctly mentions this fact on the following page. Speaking of the third and fourth month (page 911) he says: "The walls of the right ventricle, although thinner in the beginning than those of the left, soon attain the same thickness as these [of the left ventricle], which relation also remains during the whole future embryological time." It seems to me we should expect this *a priori* when we remember that the free communication of the blood, before entering the ventricles through the foramen ovale, and afterwards through the ductus arteriosus, permits the equalization of blood and work for the two ventricles. In the absence of any more satisfactory explanation, Peacock is disposed to think that the more immediate connection of the right ventricle with the circulation in the umbilical arteries, may explain the greater liability to disease of the pulmonic orifice: for the circulation in the cord and the placenta would appear to be more liable to temporary obstruction than that in the body of the fœtus itself. He says: "We know that in after-life the variable pressure of the blood in the arterial system is a fruitful cause of disease in the aortic valves." But it is just as probable that a temporary obstruction of the umbilical arteries will prevail on the valves of the aorta as on those of the pulmonary artery. The pulmonary artery may supply the greater part of blood in the descending aorta and umbilical arteries, but it does not necessarily follow that variation of pressure in the latter will cause this variation to go back, taking the longer and more indirect way through the ductus arteriosus to the valves of the pulmonary artery, more quickly than to the aortic valves. What has been claimed for the ductus arteriosus before may also answer here: this ductus will permit the equalization of pressure in the two arteries very readily.

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[1] See Kussmaul. Zeitschr. f. ration. med., loc. cit.

Returning to our own case, the theory of primary obstruction of the pulmonary artery still offers the most plausible explanation for the condition of this heart. The product of an inflammation, which may have caused the atresia, may have disappeared in the rapid progress of development after the end of the second month of intra-uterine life. The granular nodule, however, on the mitral valve goes to show that this heart was certainly not free from inflammation. The large foramen ovale permitted the surplus blood from the right heart to enter the left auricle, preventing an extra pressure on the walls of the right ventricle during their diastole and thus did not cause a dilation of this ventricle. As soon as the right ventricle contracted, and the valves of the right auriculo-ventricular aperture were closed, it took strong pressure on the part of the right ventricle to press the blood through its only small outlet, the aperture of the septum ventriculorum, and to overcome the pressure of the left ventricle. This explains the concentric hypertrophy of the right ventricle.

If we accept the theory advocated by Louis, Stillé, Rokitansky, and Peacock, that cyanosis is caused by congestion of the venous system, then we can readily account for the fact that no cyanosis was noticed during life or even directly before death in our case. It cannot be doubted that a free intermixture of blood did take place in this case; and the large foramen ovale may have prevented a stasis in the venous system. The condition of the kidneys appears to contradict this, as the hypertrophy of the connective tissue can be best explained by a venous congestion. It is highly probable that there was a venous congestion during intra-uterine life, which, I think, can be accounted for. As soon as the pulmonic orifice was closed the aorta was the only outlet for the blood of both ventricles. Now, if the compensatory dilatation of the aorta did not proceed as rapidly as the closure of the pulmonary artery, a venous stasis necessarily followed. This venous congestion could cause an exuberant growth of the connective tissue of the kidneys, which may have prevented their proper development after the aorta was wide enough to permit the free passage of the blood from the venous system, and caused them to be very small at the time of death.

The references to the principal papers on congenital atresia of the pulmonary artery will be found in the following :—

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ARTICLE IV.

RAPID LITHOTRITY WITH EVACUATION.

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THIS operation, to which its inventor has given the name Litholapaxy, has been sufficiently long before the profession to be tested and to be declared a successful procedure. It seems to be upon the eve of general